# RADIOLOGIC CASE STUDY

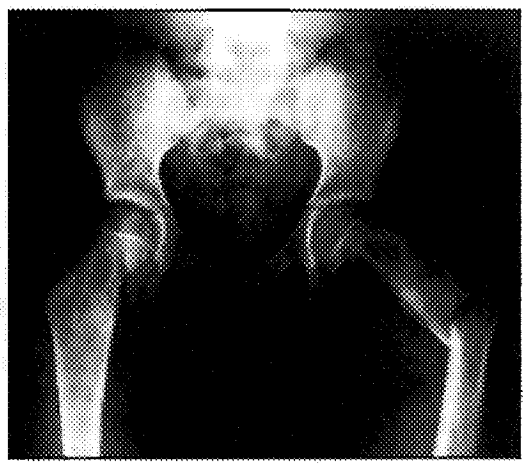

**Fig.**

A 10-year-old boy presented with acute onset of right hip pain after minor trauma. The patient gave no history of skeletal pain prior to the accident. The patient has a long history of a mild limp. On physical examination, he was noted to have a mild leg length discrepancy and several pigmented lesions with serpentine borders on his back, neck, and buttocks. The family history was noncontributory.

Radiographs of the hips demonstrate bilateral, asymmetric, expanding, lytic lesions in the pelvis and both proximal femurs (Fig). A pathologic fracture of the right femur is noted through an area of sclerosis. *Your diagnosis is?*

# POLYOSTOTIC FIBROUS DYSPLASIA

*Eddy Lucas, MD*
*Murali Sundaram, MD*
*Thomas Boccini, MD*

The pathologic fracture is through a benign appearing lesion, and there are other lesions suggestive of disseminated bone disease that raise a differential diagnosis.

The appearance of the lesions is not of a malignant process, as would be expected with metastases (unusual in this age), lymphoma, or leukemia. Other diseases with multifocal skeletal abnormalities are Langerhans' cell histiocytosis (histiocytosis X), Ollier's disease, hemangiomatosis, and brown tumors of hyperparathyroidism. The clinical clues to the correct diagnosis of polyostotic fibrous dysplasia are the leg length discrepancy and pigmented cutaneous lesions.

The radiographic pattern is not of Ollier's disease, which normally has linear islands of cartilage. Brown tumor, although plausible, is unlikely in the complete absence of any signs of rickets, which would be expected in a patient with chronic renal failure, while primary hyperparathyroidism is a very unusual diagnosis in the young. Hemangiomatosis of bone is an uncommon condition, can affect both cancellous and cortical bone, and usually presents as multiple small osteolytic foci.

## DISCUSSION

Polyostotic fibrous dysplasia is a relatively common disorder, with about 40% of all patients developing pathologic fractures.[1] There are three basic forms of fibrous dysplasia: monostotic fibrous dysplasia, polyostotic fibrous dysplasia, or either of the above associated with extraskeletal manifestations. Cafe-au-lait spots and endocrinopathies are common extraskeletal manifestations of fibrous dysplasia, with sexual precocity being the most frequent of these. Although eosinophilic granuloma may present in a similar manner, it is less likely to have as extensive involvement as in this case. In addition, cafe-au-lait spots are associated with fibrous dysplasia or neurofibromatosis and have no relationship with eosinophilic granuloma.

Endocrinopathies are present in 3% of all patients with fibrous dysplasia and may develop before or after the appearance of bony lesions. There appears to be no sex predilection for fibrous dysplasia except when sexual precocity is present, and then it has a 95% predilection for females. The role that hormonal production plays in the osseous lesions has not been established, but reactivation of osseous lesions has occurred during pregnancy and with estrogen therapy. Other endocrinopathies including hyperthyroidism, acromegaly, hyperparathyroidism, hypophosphatemic rickets, extrainsular hypothalamic diabetes mellitus, and Cushing's syndrome also may occur. The occurrence of acromegalic facies, hyperparathyroidism, or rickets may complicate the radiologic diagnosis. It also is apparent that hormonal effects may cause advanced skeletal maturation that often occurs by 5 years of age. The advanced maturation is often asymmetric and may or may not be associated with early closure of the physes.

Fibrous dysplasia accounts for 7% of all benign bone lesions and is the most common benign lesion affecting the ribs.[1] The most common sites of involvement are the ribs, femur, tibia, and humerus. Involvement of the hands and spine is uncommon. Craniofacial involvement affects half of the patients with polyostotic disease and 10% of those with monostotic disease, and may cause severe facial deformities or cranial nerve compression. Computed tomography is an excellent examination

*From the Department of Radiology, St Louis University Medical Center, St Louis, Mo.*
*Reprint requests: Murali Sundaram, MD, Dept of Radiology, St Louis University Med Ctr, 3635 Vista at Grand, St Louis, MO 63110-0250.*

to evaluate the craniofacial involvement. Rare involvement of the axial skeleton is more frequently seen with the polyostotic form and is exceptional in monostotic fibrous dysplasia.

Polyostotic fibrous dysplasia accounts for 30% of all cases and usually presents between the ages of 3 and 10 years, which is slightly earlier than that associated with the monostotic form. The earlier presentation of the polyostotic form is due to the increased frequency of symptoms and severity of the disease. There is an increased incidence of extraskeletal manifestations when compared to the monostotic form. Precocious puberty occurs in almost 50% of females, and one third of all patients with the polyostotic form have cafe-au-lait spots. The incidence of fracture is over 40% in the polyostotic form. Leg length discrepancy, shepherd's crook deformity, and craniofacial involvement is much more frequent than that associated with the monostotic form. The polyostotic form occasionally progresses throughout life, but usually becomes quiescent at puberty, as is the case for monostotic fibrous dysplasia. There are reports of severe disease causing death. The polyostotic form has a greater predilection for involving the axial skeleton and head compared to the monostotic form. The disease is asymmetric and usually bilateral, but occasionally unilateral, often affecting only a single limb; however, severe disease affecting nearly every bone is possible. The clinical course is unpredictable, but generally the earlier the onset the more progressive the disease.

Although the skeletal lesions present in a variety of patterns, their features are usually sufficient to assure the diagnosis. The radiographic features include a variety of patterns that may be lucent, sclerotic, mixed, or ground glass, with a small percentage showing calcification. Variations in bone density do not depend on the age of the lesion, but only on the relative balance of the fibrous and osseous components within the lesion. In radiolucent lesions, the cortex is usually thinned, but may be thickened after a pathologic fracture heals. When the density is increased, the cortical-medullary definition may be lost. Bony expansion is usually associated with radiolucent lesions, but does occur with dense lesions. Bony expansion most commonly occurs in the long bones, ribs, and skull. The ribs have a predilection of lucent expanding lesions posteriorly and dense sclerotic lesions anteriorly. The metaphysis is the most common site and often extends into the diaphysis, with rare involvement of the epiphysis. In the proximal femur, the lesion usually involves the intertrochanteric region, but may be confined to the femoral neck. Pelvic involvement without ipsilateral involvement of the proximal femur is rare.[2]

Fibrous dysplasia is one of several benign, bony lesions with the potential to undergo malignant degeneration. Osteosarcomas are the most common malignancy, followed by fibrosarcoma and then chondrosarcoma.[3] Degeneration into a chondrosarcoma is often difficult to recognize radiographically, as cartilaginous islands normally occur in 10% of patients with fibrous dysplasia. The incidence of malignant degeneration is higher in the patients with Albright's syndrome (4%) than in those with monostotic or polyostotic disease (0.5%). The sarcomatous degeneration usually develops during the patient's third or fourth decade of life. In approximately one third of the patients with malignant degeneration, the fibrous dysplasia had previously been treated with radiation therapy, a practice that is no longer carried out.

Surgery is currently the only acceptable treatment when function is threatened. Partial excision of the lesion followed by autologous bone grafting is now replacing the previously performed radical excision.

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